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Ancient Schwannoma of Nasal Cavity: An uncommon morphology at an unusual site

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Abstract

Introduction: Schwannoma is a benign tumour arises from the Schwann cells. Involvement of sinonasal tract is very rare constituting about 4%. In sinonasal tract ethmoid sinus is most commonly involved followed by the maxillary sinus.

Case description: We report a rare case of ancient schwannoma in a 67-year-old male, arising from the posterior wall of left nasal septum. Histopathology after complete excision of the mass confirmed the diagnosis of ancient schwannoma. IHC for S-100 showed diffuse and strong positivity.

Discussion: Common symptoms include nasal obstruction, epistaxis, anosmia, and nasal deformity due to expansile nature of the tumour. Microscopic features show hypercellular Antoni A and hypocellular Antoni B areas, and the characteristic feature of the palisading cell arrangement called Verocay bodies. Ancient schwannomas exhibit bizarre cells and extensive hyalinization, which attributes to degenerative changes.

Conclusion: Schwannomas arising from the nasal septum are very rare; however, they should be considered as differential diagnosis in patients presenting with nasal septal masses. These tumours are generally benign and show a very low rate of recurrence after excision. Management involves surgical removal followed by careful follow-up.

Keywords: Sinonasal tract, Schwannoma, immunohistochemistry

Introduction

Schwannoma is a benign neurogenic tumour. It originates from Schwann cells in the sheath of myelinated nerves. Most commonly occurs in head and neck region constituting about 25-45%, it can occur anywhere on the body, however only 4% of lesions occurs the sinonasal tract. In the sinonasal tract the ethmoid sinus is the most frequently involved site, followed in order by the maxillary sinus, nasal cavity, sphenoid sinus, and frontal sinus. In 1951, Ackerman and Taylor introduced the term “ancient schwannoma” [1].

Additionally, it tends to grow slowly. Most schwannoma cases are asymptomatic. When symptoms such as pain or neurological deficits do occur, they are usually due to compression effects on adjacent structures [2].

These symptoms include rhinorrhoea, anosmia, epistaxis and facial swelling. Patients with sinonasal schwannomas have been reported across a wide age range of 12 to 76 years, with the majority of cases occurring between 25 and 55 years of age. There is no sex predilection, as males and females are affected in equal proportions [3].

We herein report a case of a patient in 6th decade of life presenting with nasal obstruction and difficulty in breathing.

Case Report

A 67years old male presented with nasal obstruction and history of difficulty in breathing since 3years. Contrast-Enhanced Computerized Tomography (CECT) of paranasal sinuses showed a well-defined non enhancing soft tissue density polypoidal lesion in the ethmoidal sinus measuring 2.5x1.8x2.2cms, epicentered in left posterior nasal cavity at the level of middle meatus causing mild displacement of bony nasal septum towards right. Nasal endoscopy done showed a polypoidal mass attached to the posterior end of the septum (Figure 1). Polypoidal lesion was surgically excised and sent for histopathological examination. Microscopically (Figure 2, 3,4) lesional tissue showed lining of respiratory epithelium. Subepithelium showed a well circumscribed lesion comprised of hypercellular (Antoni A) areas and hypocellular (Antoni B) areas. Cells were with elongated, wavy, tapered ends nuclei and ill-defined cytoplasm interspersed with collagen fibres. Verocay bodies and numerous bizarre cells were seen. Hyalinized blood vessels, areas of haemorrhage and scattered mast cells (Figure 5) were noted. On Immunohistochemistry (IHC) S-100 (Figure 6) showed diffuse positivity. Final diagnosis of Ancient schwannoma was given.

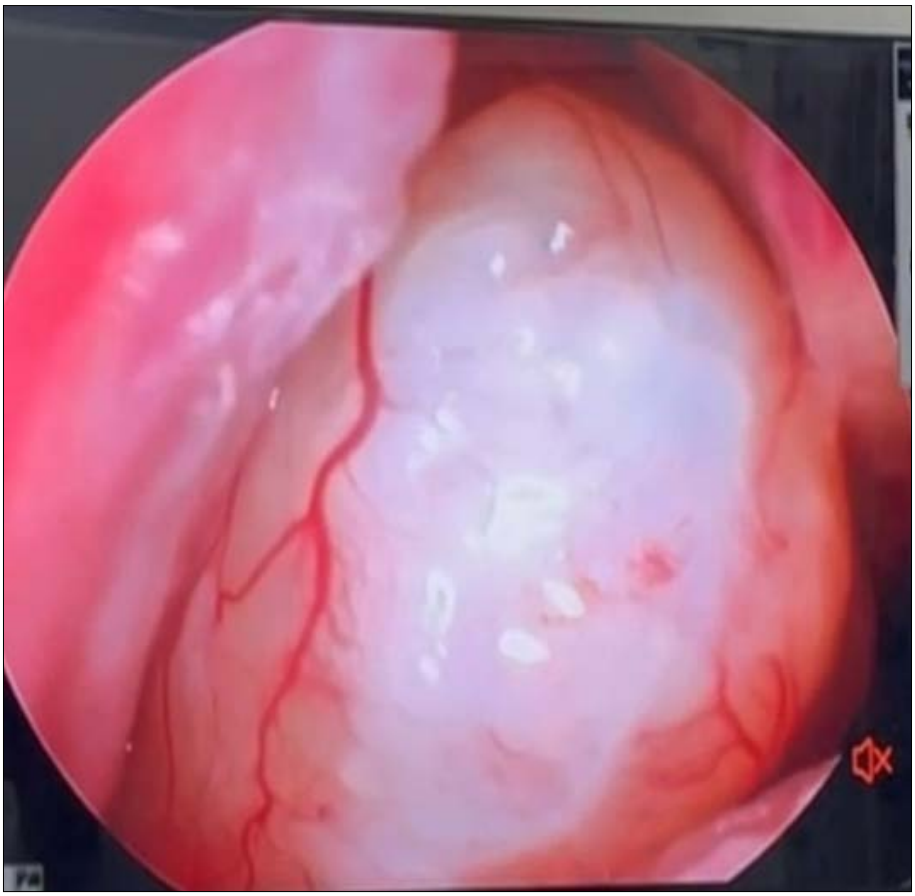

Fig 1: Nasal Endoscopy: A polypoidal mass attached to the posterior end of the septum

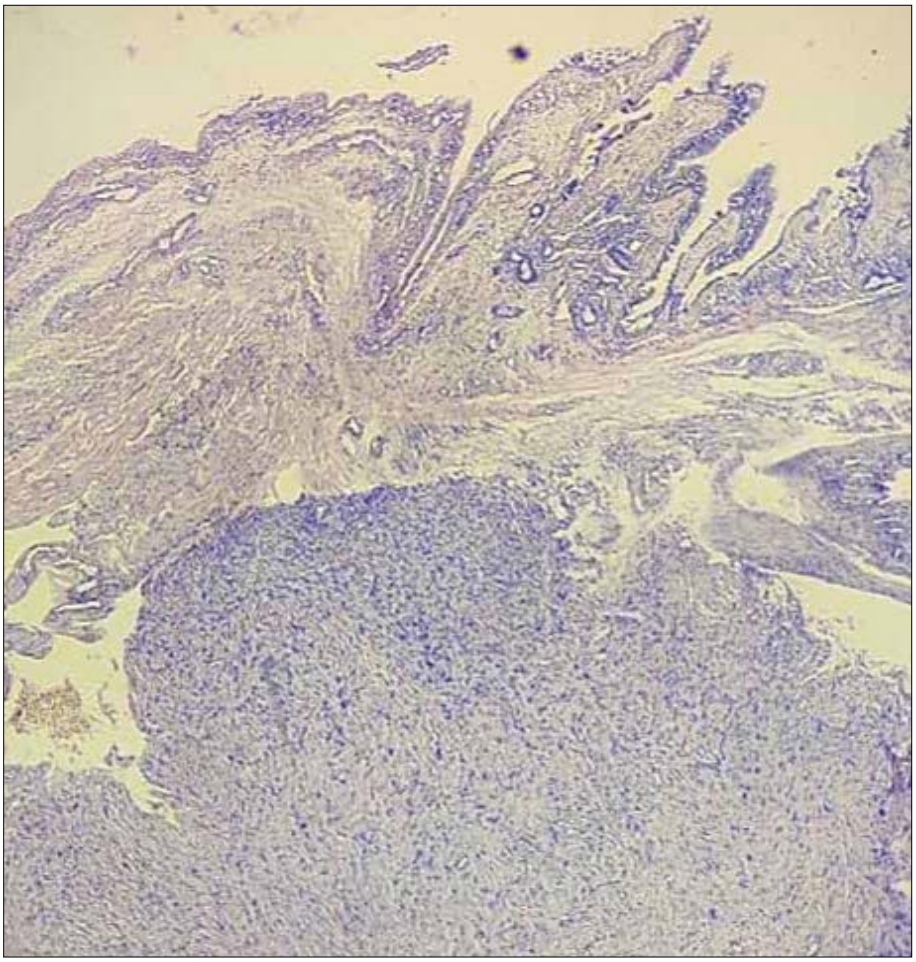

Fig 2: (H&E, 10X): Lining of respiratory epithelium (Orange arrow) and well circumscribed lesion in subepithelium (Black arrow)

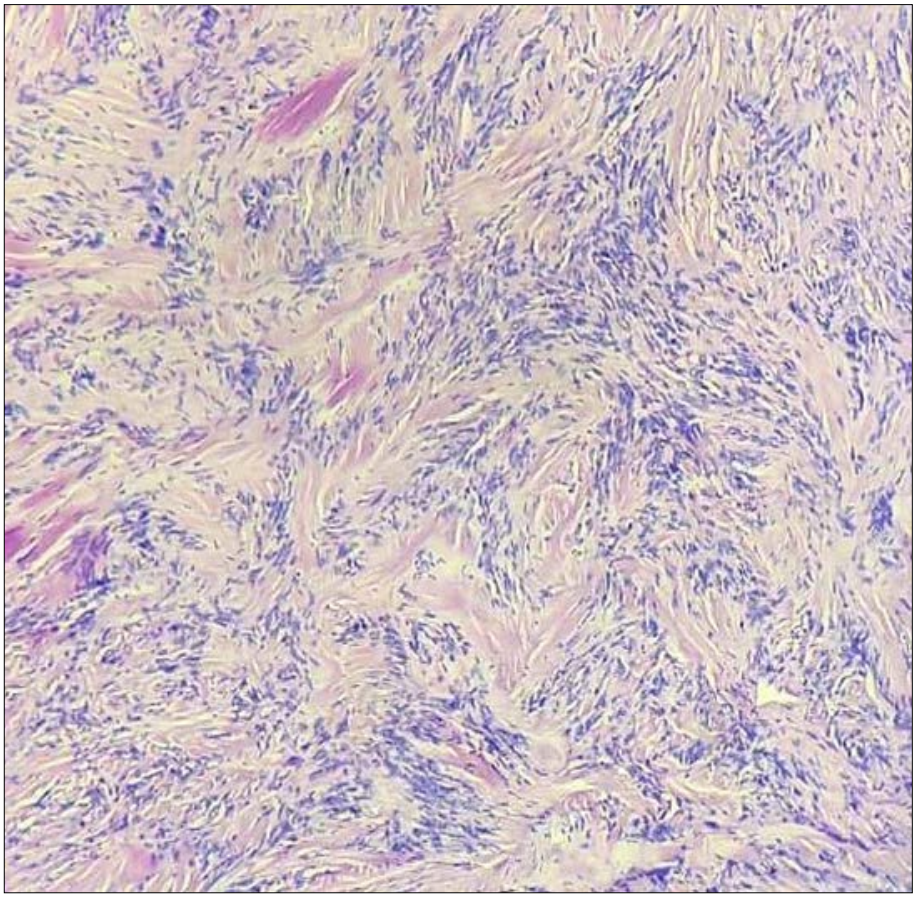

Fig 3: (H&E, 40X): Verocay bodies in Antoni A areas

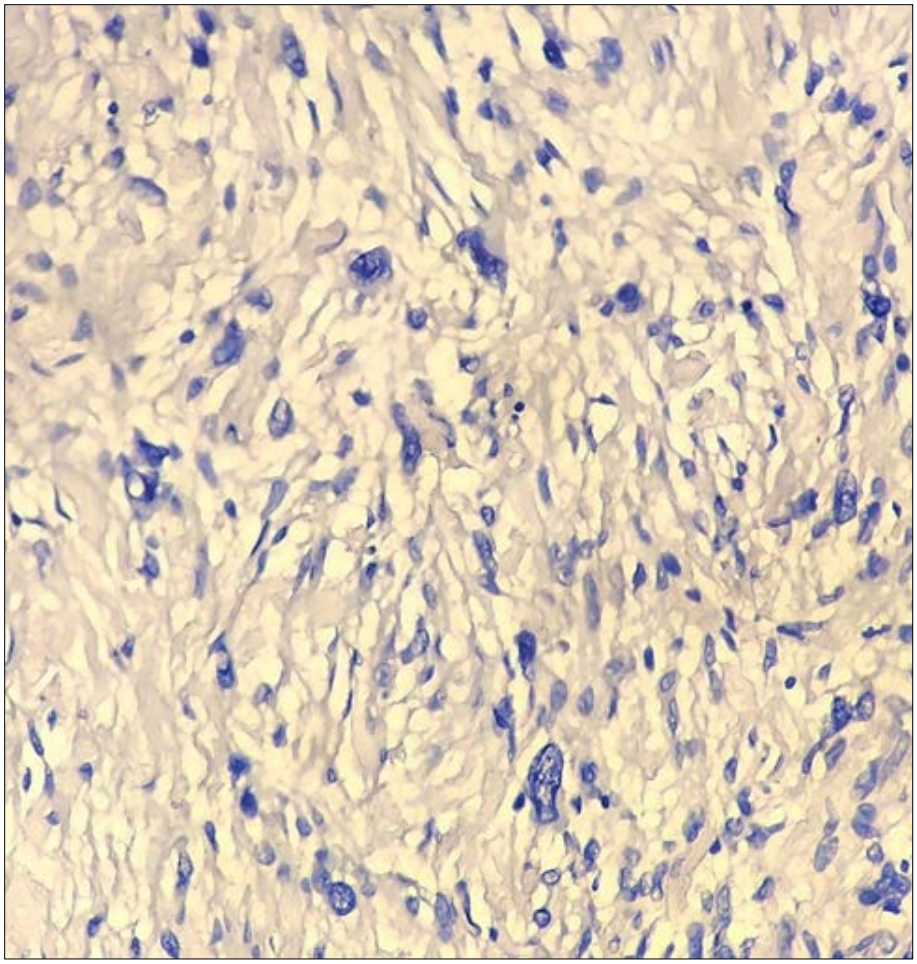

Fig 4: (H&E, 40X): Bizarre cells are seen (black arrow)

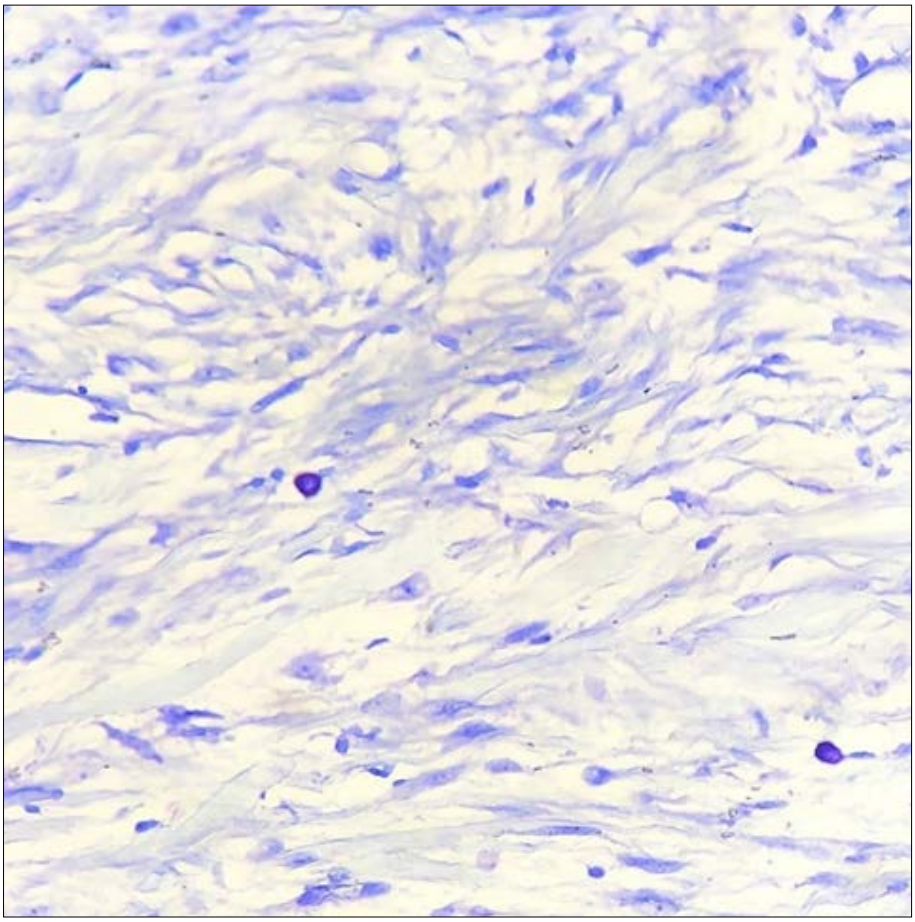

Fig 5: (Toludine blue stain, 40X): Mast cells

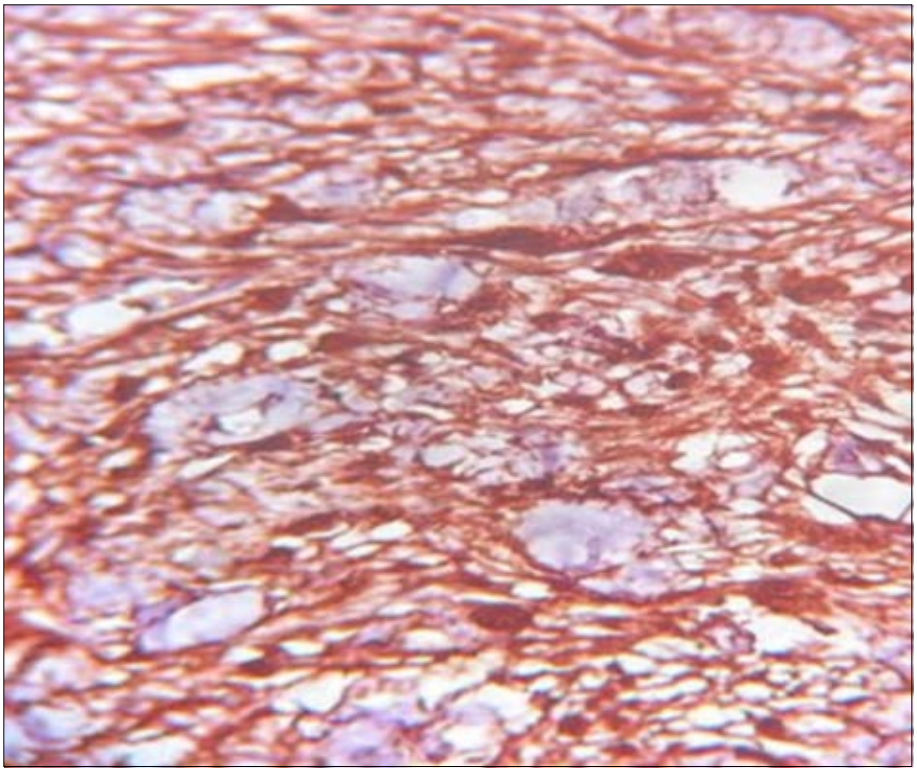

Fig 6: (IHC S-100, 40X): Shows diffuse positivity

Discussion

Schwannomas are primary tumours of the peripheral nerve sheath that originate from Schwann cells. Approximately 25–45% of schwannomas occur in the head and neck region, while involvement of the nasal cavity and paranasal sinuses is uncommon, accounting for less than 4% of cases. Current understanding suggests that sinonasal schwannomas arise from the ophthalmic and maxillary divisions of the

trigeminal nerve, or from autonomic nerves supplying the septal vessels and mucosa. The nasal septum is the most frequently reported site for sinonasal schwannomas, indicating that autonomic nerves associated with the septal vessels and mucosa are more commonly involved [4].

Schwannomas typically present in the fifth or sixth decade of life and affect males and females equally. Due to its wide range of differential diagnoses and non-specific clinical presentation, sinonasal schwannoma is difficult to diagnose on clinical grounds alone. The most common symptoms include anosmia, nasal discharge, nasal obstruction, and epistaxis. The differential diagnosis for schwannoma includes several conditions such as mucocele, inflammatory polyps, papilloma, glioma, angiofibroma, meningioma, sarcoma, squamous cell carcinoma, adenocarcinoma, melanoma, neuroblastoma and lymphomas [5].

Endoscopic features of sinonasal schwannoma are not specific; while endoscopy is useful for assessing the tumour's origin and extent, cross-sectional imaging findings are also non-specific. Therefore, histopathological examination remains the gold standard for establishing a definitive diagnosis. Microscopically Schwannomas are composed of spindle cells with two histologically distinct patterns: hypercellular Antoni type A and Antoni type B with hypocellular areas. Immunohistochemistry shows strong and diffuse positivity of the cytoplasm and nuclei for S-100 protein, a neural-crest antigen of the nervous system's supporting cells that is unique to Schwann cells and melanocytes [6].

Various histological subtypes of schwannomas are seen such as cellular, ancient, plexiform, epithelioid, microcystic schwannoma. Ancient schwannoma differs from conventional type by the presence of scattered bizarre appearing nuclei, a feature is often considered degenerative [7].

Due to the presence of hypercellularity and atypia, misdiagnosis of these lesions as sarcomas might arise. Hemangiopericytomas, leiomyomas, malignant melanomas and leiomyosarcomas these tumours often resemble schwannomas. However, the important feature which differentiates a benign ancient schwannoma from malignancy is the absence of mitotic activity. Also, the presence of a thick-walled vascular structures, capsule and areas of degenerative changes also suggest the benign nature [8].

In terms of rate of growth and malignant transformation ancient schwannoma shows similar behaviour to other types of schwannoma [9]. The preferred option for treatment of nasal septal schwannoma is surgical excision, which however depends on size and extent of the tumor. Rate of recurrence is very low [10].

Conclusion

Even though rare Schwannomas should be considered as one of the differential diagnoses in case of benign-looking mass in the nasal cavity. Histopathology is essential for confirming the diagnosis, and complete surgical excision is the treatment of choice. In summary, the presence of a schwannoma showing degenerative changes on histopathological examination, together with S100 positivity on immunohistochemistry of the nasal mass, supported the diagnosis of ancient schwannoma. Its expansile behaviour was demonstrated by compression of adjacent structures, including the nasal septum and orbital wall. Therefore, a clear understanding of the clinical features, diagnostic methods, histopathological findings, and management strategies of sinonasal schwannomas is important to ensure optimal patient outcomes and reduce the likelihood of recurrence**.**

Conflict of Interest: Nil

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